

*AMENDMENTS TO THE SPECIFICATION*

*Please replace paragraph [0025] with the following paragraph:*

[0025] Figures 12A-12I. *HERG* point mutations identified in three LQT kindreds. Pedigree structure of K1956 (Figure 12A), K2596 (Figure 12C) and K2015 (Figure 12E) are shown. Below each pedigree, the results of SSCP analyses with primer pair 5-11 (K1956) (Figure 12B), primer pair 1-9 (K2596) (Figure 12D) and primer pair 4-12 (K2015) (Figure 12F) are shown. Aberrant SSCP conformers cosegregate with the disease in each kindred. DNA sequence analyses of the normal and aberrant conformers reveals a C to T substitution at position 1682 in K1956. This mutation results in substitution of valine for a highly conserved alanine residue at codon 561 (A561V) (Figure 12G). Analyses of K2596 reveals an A to G substitution at position 1408 (T to C substitution on the anti-sense strand is shown) (Figure 12D). This mutation results in substitution of aspartic acid for a conserved asparagine in the second transmembrane domain (N470D) (Figure 12H). Analyses of K2015 reveals a G to C substitution (C to G substitution on the anti-sense strand is shown) (Figure 12F). This mutation occurs in the splice-donor sequence of intron III (see Curran et al., 1995) (intron 9 here) (Figure 12I; SEQ ID NO:43). Coding sequences are upper case and intronic sequences are lower case. Note that the G to C substitution disrupts the splice-donor site. (*HERG*, M-eag, *elk*, Warmke and Ganetzky, 1994; R-eag; Ludwig et al., 1994).